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A publication of Thomas Jefferson University, Department of Neurological Surgery

Developing Clinical Guidelines

- Assessing ESI Variations
- Minimally Invasive Skull Base Surgery
- CASE REPORTS: Hemangioblastoma
- Cervical Intramedullary Ganglioglioma
- Metastatic Spinal Tumor

Robert H. Rosenwasser, M.D., FACS, FAHA

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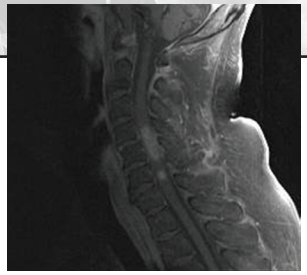

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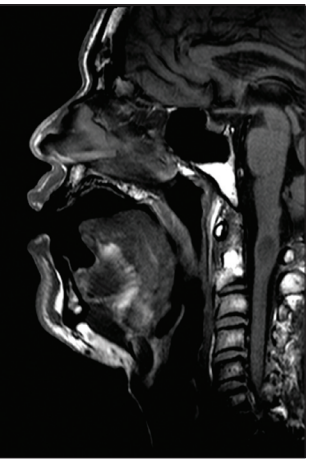

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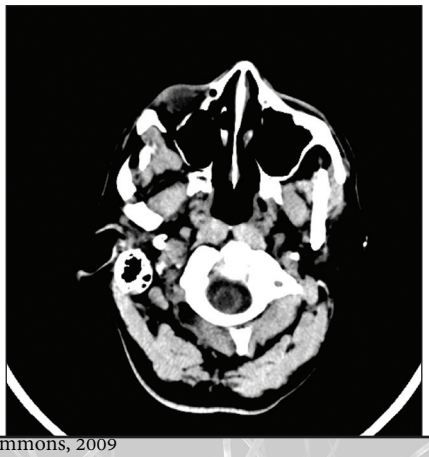

Minimally Invasive Surgery for Skull Base Tumors

James J. Evans, MD[1] and Marc R. Rosen, MD[2]

[1]Department of Neurological Surgery and [2]Department of Otolaryngology-Head and Neck Surgery Thomas Jefferson University, Philadelphia, Pennsylvania

The Jefferson Center for Minimally Invasive Cranial Base Surgery and Endoscopic Neurosurgery reflects three of the current evolutions in neurological surgery. The first of these is reflected in the name of the Center itself. Surgical Procedures, Minimally Invasive, a Medline Subject Heading since 1998, is defined as:

> **Procedures that avoid use of open invasive surgery in favor of closed or local surgery. These generally involve use of laparoscopic devices and remote-control manipulation of instruments with indirect observation of the surgical field through an endoscope or similar device. With the reduced trauma associated with minimally invasive surgery, long hospital stays may be reduced with increased rates of short stay or day surgery.**

Traditionally, cranial base tumors have been removed by making craniotomies or cranial base ostomies, and possibly by removing facial bones. To access these areas, surgeons usually need to make potentially disfiguring incisions in the face and scalp. Sometimes the morbidity from the "open" cranial base approach alone could be significant, even with an uneventful removal of the tumor.

At the Center, the endoscopic approaches are usually through the nose or nasal passages (Figure 1), however transoral endoscopic approaches to the cranial base and cervical spine are also performed. Because morbidity from the minimally invasive endoscopic approaches is so low, it becomes possible to treat patients with tumors that were previously considered non-resectable or as having too poor a prognosis for more invasive surgery. Even partial resection of such tumors can relieve pain, preserve function, and permit earlier adjuvant radiation and chemotherapy.

James Evans, MD and Marc Rosen, MD, both faculty at Jefferson Medical College of Thomas Jefferson University (Evans in Neurological Surgery, Rosen in Otolaryngology-Head and Neck Surgery), are the Center's co-directors. Their collaboration represents another evolution, the increasing interaction between medical and surgical disciplines (Figure 2). Both specialties bring unique instruments, surgical techniques, and clinical experience that have been merged to exponentially increase our understanding and management of cranial base tumors and disorders. Treatment of these patients in the Center also draws upon knowledge and resources from many other departments including Radiology, Oncology, and Ophthalmology.

The third evolution is in the tracking of patient outcomes. Through the Center, we have developed a specialized database for tracking patient demographics, neurological examinations, ENT parameters, endocrine function, ophthalmologic evaluations, tumor control, morbidity, and outcomes. Data collection is essential for confirmation of treatment efficacy and safety. Continual re-evaluation of the treatment process and results can be critical when developing and validating new endoscopic techniques. Also, the "Pay for Performance" movement is a recent addition to the reasons why clinical organizations need to collect and store data that can be searched and cross-referenced. Data at our Center is stored and retrieved using a Microsoft Access database application designed in-house (Figures 3 and 4). With five years of accumulated data, the Center is now getting a clear view of what it has achieved and what the next goals should be.

Many of the Center's patients present with pituitary tumors. Pituitary adenomas represent one of the most common brain tumors and were the first type of tumor where endonasal endoscopic techniques were used. Sixty percent of the patients seen by the Center for such tumors present with visual deterioration. A recent study published by members of our Center has shown that ophthalmologic studies such as optical coherence tomography not only document the extent of presenting visual loss, but may be used as a predictor of visual improvement with resection of the tumor and decompression of the optic chiasm.

Table 1. Advantages of Endoscopic Surgery over "Open" Surgery

Better preservation of function and appearance
Lower morbidity rates
Shorter hospital stays
Greater resections of tumors
Facilitates re-operations
Permits earlier post-operative radiation and chemotherapy

Another area of interest in our Center is the extension of cranial base approaches to include tumors involving the cervical spine. Figure 1 shows an endoscopic transnasal view of the posterior pharyngeal wall and accompanying intraoperative navigation images. We have recently completed a morphometric study on over 100 patients to determine the factors that affect the extent of cervical spine access via an endonasal surgical approach. These findings have led to the development of new techniques and instruments for endonasal cervical spine surgery.

Endoscopic transnasal procedures continue to benefit from technological developments such as real-time intraoperative imaging (MRI, CT), dedicated endoscopic surgical suites, and the development of new materials and techniques for cranial base repair. We are also exploring the adaptation of surgical robotics systems to augment the precise and delicate nature of these endonasal surgical procedures.

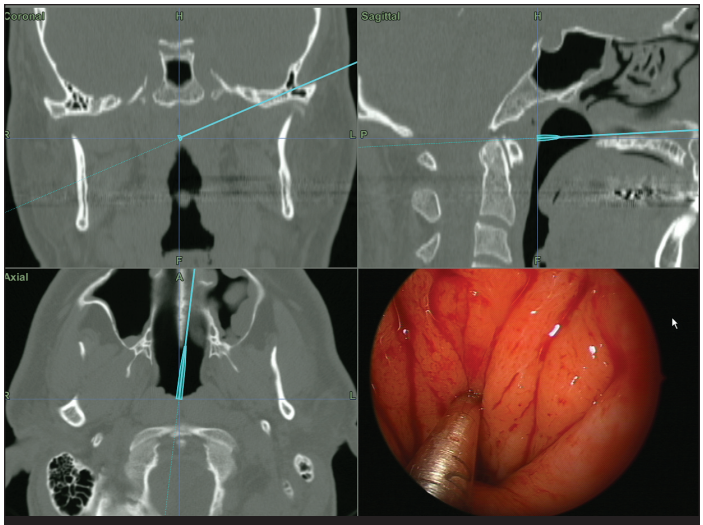

Figure 1
Endonasal navigation during surgery

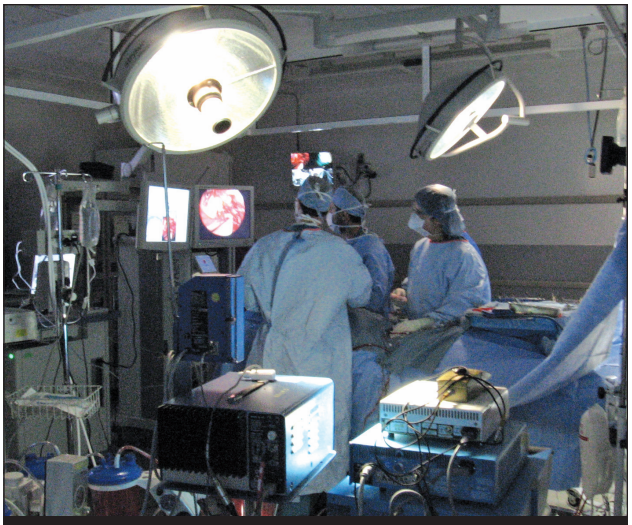

Figure 2
Multidisciplinary surgical team

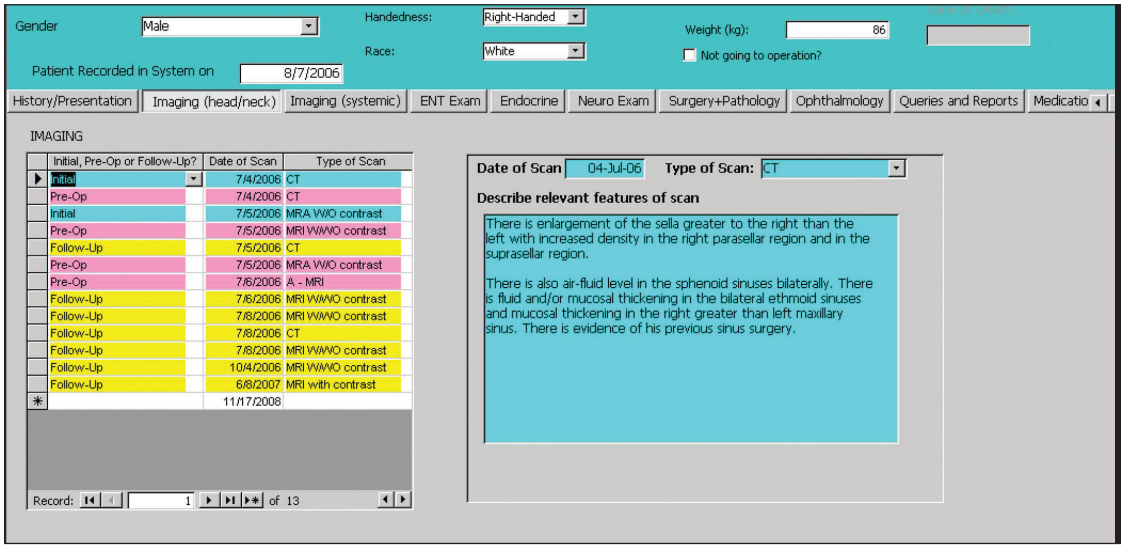

Figure 3
Interface page for database – image records are color coded by type, and selecting image record on left updates notes field on right

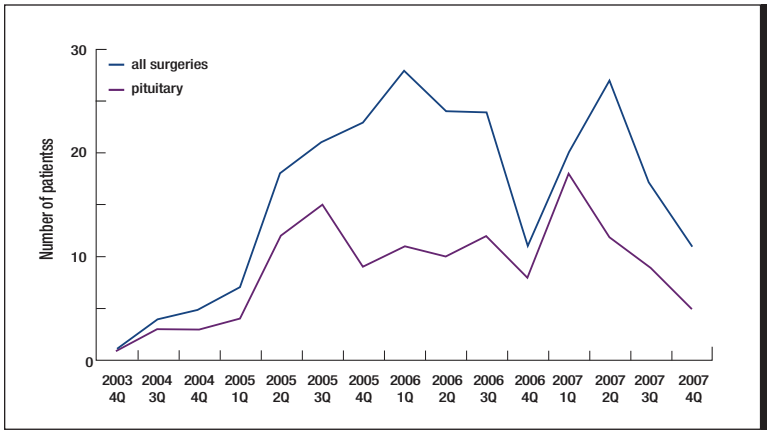

Figure 4
number of surgical cases, by quarter, through end of 2007

Clinical Guidelines Written by Residents

David W. Andrews, MD

Department of Neurological Surgery, Thomas Jefferson University, Philadelphia, Pennsylvania

"Variation" is an innocent word that that can represent many levels of frustration to the clinician. Variation among patients is the least of these; the physician expects patients and their individual problems to be as diverse as the human race itself. Variation within a practice should be due to matching the specific needs of the specific patient. Other variations can mean trouble if they represent differences in understanding of the problem among clinicians and other allied health practitioners. These differences could be between institutions or even between shifts within one institution.

The Tufts Health Care Institute (THCI) is a non-profit organization which provides educational tools for medical and surgical residents across the US. One of these tools teaches clinical practice guidelines, defined by the Institute of Medicine as "systematically developed statements to assist practitioner and patient decisions about appropriate health care for specific clinical circumstances."[1]. One of the authors of the Clinical Guidelines module is David B. Nash, Professor of Health Policy here at Thomas Jefferson University.

The Jefferson Department of Neurological Surgery has been participating in the THCI program for the past few years, mentored by Dr. David W. Andrews of the department. Because surgery and medicine have different emphases, Dr. Andrews has found that some of the THCI modules are more appropriate than others for the neurosurgical residents, and that the Clinical Guidelines module in particular has generated both enthusiasm and good results. A selection of the TJU neurosurgical resident-generated guidelines are presented here. These guidelines have not officially been adopted by the department or by any institution that we know of, but show how clinical problems can be identified and solved.

References

1. (Institute of Medicine, 1990, p. 38) Institute of Medicine. Field MJ, Lohr KN (eds.) Clinical Practice Guidelines: Directions for a New Program, page 38. Committee to Advise the Public Health Service on

Acknowledgement

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Anticonvulsant use in brain tumor patients

Hugh Moulding, MD

More than 200,000 patients are diagnosed with primary or metastatic brain tumors each year in the United States. Of these patients, 20% to 40% will develop seizures at presentation, and another 20% to 40% will require treatment for seizures during their illness. Although the use of antiepileptic drugs (AEDs) in patients who have had seizures seems reasonable, the issue of prophylactic AED use for patients who have not had a seizure is an intensely debated subject.

At TJUH, we see the wide variety of practices in prophylactic use of anticonvulsants. Some physicians will use anticonvulsants and follow therapeutic levels, others will use a starting dose of AED and not follow levels, and still others will not use AED at all. The reason for the discrepancy is most likely multifactorial. There is little evidence to the benefit of AEDs in patients who have not had seizures when considering the side effect profile of AEDs. However, give the litigious climate of Philadelphia, many neurosurgeons are reluctant to leave a patient with an intracerebral lesion without antiepileptic coverage. Additionally, many neurosurgeons are creatures of habit, trained to practice a certain way, and have done so for many years during which "it has worked or them so far." A set of guidelines based on a literature search, where level I evidence is cited where possible, would help physicians to be more comfortable about changing their practice.

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Since 2000, there have been practive guidelines in place approved by the American Academy of Neurology (see Ref 3) based on peer reviewed publications with level I evidence. Despite these practice parameters a recent survey reported that 81% of neurosurgeons and 53% of neurologists prescribed antiepileptic drugs (AEDs) prophylactically.

For the 20-40% of patients diagnosed with a brain tumor who have experienced a seizure, prophylactic use of AEDs is universally accepted. Seventy percent of brain tumor patients presenting with seizures will suffer recurrent seizures regardless of tumor type. However, patients that have not had a seizure still remain at risk and 20-45% will develop seizures. Some factors that have been shown to increase the likelihood of developing seizures are:

1. Tumor location, most likely in frontal, parietal or temporal lobes, especially parasagital meningiomas (74% seizure prevalence).
2. Number of tumors: increased number of tumors correlates to higher seizure risk.
3. Tumor histopathology: Slow growing tumors are at increased risk.
4. Age: younger patients more likely to seize.

One factor that is conspicuously absent from the list above is AED use. Of the twelve studies of Level I & II data that examined this question, only one reported a significant difference in seizure frequency and this favored the non-AED group having a lower seizure risk. A meta-analysis performed on the four Level I studies showed an Odds Ratio of 1.09 with no statistical significance (p=0.9). Furthermore AED use is not benign; 23.8% of these patients experience side effects that warranted change or discontinuation of the AED. These side effects included rash (14%), nausea and vomiting (5%), encephalopathy (5%), and myelosuppression (3%).

References

1. Sperling MR, Ko J: Seizures and brain tumors. Seminars in Oncology. 33(3):333-41, 2006.
2. Stevens GHJ: Antiepileptic therapy in patients with central nervous system malignancies. Current Neurology and Neuroscience Reports. 6(4):311-8, 2006.
3. Glantz MJ, Cole BF, Forsyth PA, et al.: Practice parameter: anticonvulsant prophylaxis in patients with newly diagnosed brain tumors: report of the quality standards subcommittee of the American Academy of Neurology. Neurology. 54:1886-93, 2000.

Cervical spondylotic myelopathy

Amgad Hanna, MD

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Cervical spondylotic myelopathy is a common cause of major morbidity in middle-aged and elderly people. Various functional classifications are used to evaluate patients with cervical spondylosis. They are based on pure clinical data, and are used as a method of evaluation of the patient's progress during the course of the disease. None of these classifications defines criteria for the indication for surgery.

Patients with cervical myelopathy may be managed either conservatively or surgically. Conservative management includes medications (e..g, non-steroidal anti-inflammatories or corticosteroids), cervical orthotics, physical therapy, or injections. Surgery includes anterior and/or posterior decompression and/or fusion.

There remains considerable uncertainty regarding the decision to operate and the timing for surgery. This is left to the surgeon's discretion in most of the cases. Most surgeons would agree that patients with mild myelopathy should be managed conservatively and patients with severe myelopathy should be managed surgically. There is a large gray zone of moderate myelopathy where surgeons may decide about surgery or conservative management. The definition of mild, moderate, or severe myelopathy is vague in literature.

The timing of surgery is also controversial in literature. Advocates of early surgery try to stop disease progression, prevent further clinical deterioration of the patients, and prevent the risk of acute deterioration with neck injuries (central cord syndrome). Advocates of late surgery try to delay the potential complications of surgery like adjacent level disease, spinal cord injury, cerebrospinal fluid leak, infection, bleeding, and possible clinical deterioration.

We attempt to address the literature controversies regarding this topic, the severity of myelopathy, and the indications for surgery.

Online search for resources

1. National guideline clearinghouse. We searched for "cervical myelopathy". No guidelines were found for the management of cervical myelopathy. Related guidelines were found for cervical radiculopathy: Review criteria for cervical surgery for entrapment of a single nerve root. Olympia (WA): Washington State Department of Labor and Industries; 2004 June. 1p. The article states that "Cases of myelopathy should be referred for physician review".
2. The Cochrane Library. We searched for "cervical myelopathy". One article was found: Cochrane review on the role of surgery in cervical spondylotic radiculomyelopathy. Fouyas I, Statham P, Sandercock P. Spine 2002; 27 (7): 736-47
3. Medline search: www.ncbi.nlm.nih.gov. We searched for "management of cervical myelopathy"; 104 articles were found

Summary of findings

There are no current guidelines for the management of cervical spondylotic myelopathy. Several articles (1,2) discussed the prognostic factors and presented surgical outcomes. These articles presented conflicting results as far as the prognostic significance of signal changes on MRI, age, and duration of symptoms.

Other studies discussed the indications and outcomes of different surgical approaches. Again, conflicting results were found. Fouyas et al. found no significant longterm differences between surgery and conservative management[3]. Houten, Kim, Medow, Komotar, and Sekhon showed good results with surgical decompression [2, 4-7]. However, there was no comparison to conservative management.

Guidelines

The two articles selected are references 2 and 3.

Reference 2: Houten, Cooper (2003)

Full reference: Laminectomy and posterior cervical plating for multilevel cervical spondylotic myelopathy and ossification of the posterior longitudinal ligament: effects on cervical alignment, spinal cord compression, and neurological outcome. Houten J, Cooper P. Neurosurgery 2003; 52 (5): 1081-8

Results

- Multilevel cervical laminectomy and fusion for cervical spondylotic myelopathy provide minimal morbidity, excellent decompression, immediate stability, prevent kyphosis, and precludes further spondylosis.

Strengths

- Combined clinical and radiological criteria for outcome after surgery.
- Thirty months follow up.

Weaknesses

- Retrospective study.
- Small number of patients (38).
- No comparison group with conservative management.
- No consideration for bony fusion vs pseudoarthrosis.

Reference 3: Fouyas, Statham, Sandercock (2002)

Full reference: Cochrane review on the role of surgery in cervical spondylotic radiculomyelopathy. Fouyas I, Statham P, Sandercock P. Spine 2002; 27 (7): 736-47

Final Guideline

This guideline targets mainly neurosurgeons and orthopedic spine surgeons who need to decide for their patients whether to proceed with surgical or conservative management. It also targets primary care physicians to guide them as to which patients need to be referred for surgery.

Evaluation of the severity of myelopathy:

The following scoring system is proposed to evaluate the severity of cervical myelopathy:

Comments

- This is the first grading system to include both clinical and radiological data.
- Mild to moderate symptoms: mild pain or numbness. Severe symptoms: severe pain or numbness, loss of dexterity, difficulty in walking.
- Mild to moderate signs: hyper-reflexia, Hoffman's or Babinski's signs, mild weakness. Severe signs: significant weakness, spasticity.
- Moderate stenosis: Mid-sagittal diameter: 10-12 mm. Severe stenosis: Mid-sagittal diameter: < 10 mm.
- The recommendations are based on the best available literature.
- This is just a guideline. Cases need to be managed individually, based on the whole clinical scenario, progression of the disease, and co-morbidities.

Table 1. Score System

No Symptoms	0	Mild to Moderate Symptoms	1	Severe Symptoms	2
No Signs	0	Mild to Moderate Signs	1	Severe Signs	2
No stenosis	0	Moderate Stenosis	1	Severe Stenosis	2
No signal changes on MRI	0			Signal Changes on MRI	2

Total score 1-3: Conservative management. Total score 4-8: Surgery.

Results

- No significant difference in long term outcome between surgical and conservative management for cervical myelopathy.

Strengths

- Prospective randomized trials.

Weaknesses

- Patients represented in the studies had mainly mild functional deficit. They do not represent the population that usually benefits from surgery, who have moderate to severe myelopathy.

References

1. Effect of intramedullary signal changes on the surgical outcome of patients with cervical spondylotic myelopathy. Suri et al. Spine J 2003; 3 (1): 33-45
2. Laminectomy and posterior cervical plating for multilevel cervical spondylotic myelopathy and ossification of the posterior longitudinal ligament: effects on cervical alignment, spinal cord compression, and neurological outcome. Houten J, Cooper P. Neurosurgery 2003; 52 (5): 1081-8
3. Cochrane review on the role of surgery in cervical spondylotic radiculomyelopathy. Fouyas I, Statham P, Sandercock P. Spine 2002; 27 (7): 736-47
4. Indications for circumferential surgery for cervical spondylotic myelopathy. Kim P, Alexander J. Spine J 2006; 6 Suppl: S 299-307
5. Surgical management of cervical myelopathy: indications and techniques for surgical corpectomy. Medow J, Trost G, Sandin J. Spine J 2006; 6 Suppl: S 233-41
6. Surgical management of cervical myelopathy: indications and techniques for laminectomy and fusion. Komotar R, Morcco J, Kaiser M. Spine J 2006; 6 Suppl: S 252-67
7. Posterior cervical decompression and fusion for circumferential spondylotic cervical stenosis: review of 50 consecutive cases. Sekhon L. J Clin Neurosci 2006; 13 (1): 23-30

Does the treatment of carotid dissection require anticoagulation with coumadin or is antiplatelet therapy equally efficacious?

Aditya Pandey, MD

http://commons.wikimedia.org/wiki/Image:Blut-EDTA.jpg

Vessel dissection represents injury to the wall of the vessel either from the luminal side or adventitial (outside) side. Extracranial carotid and vertebral artery dissections represent the second leading cause of stroke in young adults. Its incidence is 2-3 persons/ 100,000/ year. The mechanism of developing stroke within this patient population is from clot formation at the site of the injury to the vessel. The causes of carotid dissection are numerous, but it most commonly occurs due to a traumatic injury in an individual who has weakened vessels. To prevent such an event, patients are started on blood thinners: either antiplatelet (aspirin) or coumadin.

Depending on physician and surgeon preference, different anticoagulation therapy can be used for the same type of patient with carotid

dissection. Those who use coumadin do so since they have had success in treating carotid dissection patients without stroke formation. The same is true of physicians utilizing antiplatelet therapy. The use of coumadin not only requires that patients have frequent blood tests to show level of blood thinning but also that the bleeding risks associated with it are higher. The goal of this analysis is to present the evidence for using coumadin versus aspirin in the treatment of carotid dissection.

Benefits of Full Anticoagulation

- anticoagulation leads to the prevention of stroke from the formation of clots at the site of the vessel injury

Complications of Full Anticoagulation

- anticoagulation could lead to further bleeding into the brain

Specifics of Search

- Search of National Clearinghouse Guidelines revealed one review but no specific guidelines
- Search of the Cochrane Database did not reveal any position statements on the subject
- The following key term searches on Pubmed led to numerous articles on the subject of treating carotid dissection with anticoagulation:
  - "Extracranial carotid dissection"
  - "Treatment of extracranial carotid dissection"
  - "Anticoagulation for extracranial carotid dissection"

Results

1. Antithrombotic drugs for carotid artery dissection. Cochrane Database Syst Rev. 2000; (4): CD 000255

Strengths

- comprehensive review of case controlled studies and case series
- applies to carotid dissection patients who had undergone either anticoagulation with coumadin or antiplatelet therapy

Limitations

- no randomized control trials included in this study (No RCT have been performed addressing this issue)

Summary

- 26 eligible studies were reviewed (327 patients)
- No statistically significant difference in outcome (mortality/disability) between patients on antiplatelet therapy versus those on anticoagulation (coumadin)
- No difference in the occurrence of intracerebral hemorrhage between the two groups (0% for antiplatelet group and 0.5% for anticoagulation group)

Conclusion

- no statistically significant difference in outcome or complication when comparing the anticoagulation versus the antiplatelet group of patients

2. Cervical Arterial Dissection: time for a therapeutic trial? Stroke. 2003 Dec; 34 (12): 2856 – 60

Strengths

- prospective enrolled patients with carotid and vertebral dissections
- large number of patients (n=116)
- comparison of aspirin vs. anticoagulation

Weaknesses

- not a randomized control trial
- N might not be large enough to a small statistically significant difference

Summary

- Canadian Stroke Consortium prospectively enrolled patients with extra-cranial dissections and followed patients for one year. The endpoints being evaluated included: TIA, Stroke, or Death. 105 patients had complete follow up. In patients treated with anticoagulation the event rate was 8.3% versus those treated with aspirin where the event rate was 12.4%. While there was an absolute difference of 4.1%, this difference was not statistically significant.

Conclusion

There were no statistically significant differences between the outcome rates of patients on ASA versus those on anticoagulation.

1. Dissection of Cervical Arteries: Long-term follow-up study of 130 consecutive cases. Cerebrovasc Dis. 2006; 22 (2-3): 150-4

Strengths

- Prospective Study
- N=130 cases
- Comparison of ASA vs. anticoagulation

Weaknesses

- not a randomized control trial
- F/U only 6 months

Summary

- 130 patients with angiographically proven cervical arterial dissection were followed for the events of stroke and death. There was no significant differences in outcome were found when comparing patients on aspirin versus the patients on anticoagulation.

Conclusion

In patients with proven carotid dissections, there is no difference in outcome between patients treated with ASA vs. those treated with anticoagulation.

1. Outcome of extracranial cervicocephalic arterial dissections: A follow-up study. Neurol Res. 2002 Jun; 24 (4): 395-8

Strengths

- comparison of ASA vs. anticoagulation
- long term f/u (nearly 10 years)

Weaknesses

- not a randomized control trial
- small patient population (n=27)

Summary

- 27 patients with extracrainal CAD who were treated and followed by the stroke service. Outcome was assessed using the modified Rankin Score and recurrent stroke and TIAs were also recorded. The outcome was favorable with either antiplatelet or anticoagulation.

Conclusion

Either antiplatelets or anticoagulation are equally effective in preventing strokes after carotid dissection,

Final Guideline

In individuals with a proven extracranial carotid dissection, aspirin therapy should be initiated instead of coumadin. The current literature on the subject shows no statistically significant difference in outcome of individuals treated with aspirin versus those treated with coumadin in the setting of carotid dissection. While the risks with coumadin are not higher, it still requires evaluation of the INR for accurate dosing. Such is not the requirement of antiplatelet therapy. In addition other medications (cimetidine) can affect the metabolism of coumadin thus leading to inappropriate levels of anticoagulation. Thus antiplatelet therapy is an effective method of preventing strokes in individuals with carotid dissection.

http://commons.wikimedia.org/wiki/Image:Philadelphia_Fire_Department_medic_unit.JPG

Would a standardized protocol for the inter-hospital transport of critically ill patients alter outcomes?

Jack Klem, MD

The rapid evolution of healthcare technology has created a disparity among hospitals and thus led to the establishment of highly specialized quarternary institutions, termed "Centers of Excellence." This has created a need for the systematic triage and transport of certain critically ill patients that would not receive adequate care at a community hospital. Emergency Medical Services (EMS) has made the transport of patients a national issue due to regionalization, specialization, and facility designation by payers. Despite this, there exists a paucity of guidelines to direct the appropriate flow of patients to higher levels of care.

Diversity exists amongst transport teams and overall resource allocation. Interfacility transport is provided by a variety of levels and types of personnel and agencies. The medical condition of the patient is not always matched appropriately with the acuity of care provided by the transport team. Less severely ill patients are sometimes intubated and ventilated solely to facilitate ease and safety of transport.

In addition, the disease process of a particular diagnosis is not always understood prior to dispatching the appropriate means of transport (ie, air vs. ground). For example, patients with suspected aneurysmal subarachnoid hemorrhage require the most expeditious mode of transportation due the high risk of re-rupture in the acute period. Moreover, certain diagnoses such as cerebellar hematoma with deteriorating neurological function require immediate transfer to the operating room with no need for an available ICU bed. Recognition of these "hyperacute" scenarios are critical to patient outcome.

Guidelines are necessary to standardize interfacility transport on at least the regional level. Several steps must be analyzed in order to establish a systems-based protocol for interfacility transfer. The type of transfer must be established (ie, hospital to hospital, rehab to hospital, clinic to hospital, etc.) Next, provider capabilities must match the patient's current and potential needs in order to provide safe and effective care during transport. This has been shown to impact outcomes in the transport of pediatric patients. On an administrative level, certification of necessity for transfer is a requirement for reimbursement by Medicare and Medicaid. This is directed by federal legislation outlined in the Consolidated Omnibus Budget Reconciliation Act (COBRA) and Emergency Medical Treatment and Labor Act (EMTALA). In summary, all of these factors must be integrated in order to develop a regional or even national plan for interfacility transport. This shall streamline resource allocation and potentially improve outcomes

Online Search

1. National Guidelines Clearinghouse was queried for "patient, hospital transport" and "interhospital transport"
2. Google was queried for "patient transport, guidelines" and "hospital transport"

Study #1: Warren at al (2004)

Full reference: Warren et al. Guidelines for the inter- and intrahospital transport of critically ill patients. Crit Care Med 32:1 (2004) 256- 262.

Purpose

- Development of practice guidelines for inter- and intrahospital transport of the critically ill patient in order to establish an efficient, organized process supported by appropriate equipment and personnel. This would ultimately enhance patient safety.

Methods

- Synthesis of prospective clinical outcome studies, retrospective reviews, and anecdotal reports by a task force of experts providing consensus opinion.

Results

- A multidisciplinary team of physicians, nurses, respiratory therapists, hospital administration and the local emergency medical service must justify and coordinate the process by conducting a needs assessment of the referring hospital or clinic, asking whether transport is likely to alter the management or outcome of the patient. A patient must have a preliminary diagnosis that can be further refined and ultimately treated at a center that offers specialized diagnostic capability. Receiving hospitals should then proceed with a formalized plan addressing the following points:
- pretransport coordination and communication
- transport personnel
- transport equipment:
- monitoring during transport
- documentation: The patient's medical record and relevant laboratory and radiographic studies must be copied for the accepting facility. It is also suggested to perform a COBRA/EMTALA checklist to ensure compliance with federal regulations.
- The above five points should be evaluated and refined regularly by the hospital using a standard quality improvement process. Many of the specific details from these guidelines are incorporated in the final guidelines below.

Conclusions

When services are required that exceed available resources at a particular hospital or clinic, a patient will be ideally transferred to a facility that has the necessary resources. The decision to transport a critically ill patient is based on the potential benefits (ie, higher level of technical/cognitive/procedural care) and weighed against the risks. Justification for transport must be established and several points must be addressed to ensure safe and efficient transfer.

Strengths of study

- Article provides clear, comprehensive guidelines discussed above for transporting patient within and between hospitals. Not only do these guidelines comply with federal regulations, but they also illustrate the importance of having an organized, efficient and standardized protocol that can be followed by any hospital.
- Details of transport such as appropriate equipment and personnel are described in order to ensure utmost safety for the patient being transported. Again, this is an effort to standardize the entire process in order to eliminate "system-based" mistakes. For example, there exists much variability in the acuity of care provided

by transport teams due to improper triage, insufficient equipment, and inexperienced personnel. To resolve this, this article takes the stance that patient transportation must be subjected to the same rigors as the aviation industry in eliminating systems errors by establishing a universal set of guidelines.

Weaknesses of study

- This is really the first effort in the literature to establish a universal set of guidelines and, as a result, no outcome studies are yet available.
- This article also assumes that every hospital has the same basic resources available to standardize patient transport. This becomes an issue with the transport of critically ill patients where the accepting hospital may have to provide the appropriately staffed and equipped transportation team.

Study #2: Fan et al (2006)

Full reference: Fan et al. Outcomes of interfacility critical care adult patient transport: a systematic review. Critical Care 10 (2006) 1-7.

Purpose

- Determine the adverse events and important prognostic factors associated with interfacility transport of intubated and mechanically ventilated adult patients.

Methods

- A systematic review of multiple databases yielded 5 case-series comprising of a total of 245 patients. Two of the case-series were prospective in design.

Results

- Data was synthesized in a qualitative manner due to significant heterogeneity in study population, outcome events, and results. The most common indication for interfacility transport was a need for specialized investigations and interventions. Transport modalities included air (66%), ground (31%) and commercial aircraft (3%). Transport teams included a physician in 3 of the 5 studies. Death during transport was rare (n=1) and no other adverse events or significant therapeutic interventions were reported during transport. Of note, one study reported a 19% (28/145) incidence of respiratory alkalosis upon arrival.
-

Conclusion

Insufficient data exists regarding the mortality, morbidity, and risk factors associated with the interfacility transport of critically ill patients. Further research is necessary to understand which patients are most at risk while being transported. Recognizing the types of events that can occur is an important step in patient preparation and planning.

Strengths of study

- First attempt to study outcomes of inter-hospital transport of critically ill patients

Weaknesses of study

- This is similar to a meta-analysis that combines different levels of evidence (ie, three retrospective studies plus two prospective studies) and attempts to draw meaningful conclusions. There must be one prospective trial that addresses the same patient population.
- This article looks only at adverse outcomes that occur while a patient is en route rather than considering the period just before and after transport. This under reports situations, for example, where some patients begin to deteriorate and are intubated immediately upon arrival.

Study #3: Ligtenberg et al (2005)

Full reference: Ligtenberg et al. Quality of interhospital transport of critically ill patients: a prospective audit. Critical Care 9 (2005) 446-51.

Purpose

- Determine the adverse events and important prognostic factors associated with interfacility transport.

Methods

- prospective study describing 100 consecutive ICU transfers, of which 65% were mechanically ventilated and 38% on vasoactive drugs.

Results

- Data was collected on adverse events before, during, and after transport. 34% of patients incurred adverse events with 6 deaths being reported within the first 24-hours after arrival. This study was different because not all patients were intubated and not all were accompanied by a physician as was the case in Fan et al. (2006).

Conclusions

Important factors for improvement were better overall communication between the referring/ receiving hospital and strict adherence to checklists/ published protocols.

Strengths of study

- Describes a single, prospective study involving the transfer of 100 patients into only one university center. This eliminates the inherent flaws of a combining several different studies.
- Information regarding adverse events was collected for 24-hours after arrival.

Weaknesses of study

- There was no stratification of diagnoses. For example, there is no discussion as to why each of the 34 adverse events occurred. For example, was this due to inadequate stabilization of the patient's medical condition prior to transport, due to inadequate care provided during transport, or merely patient disease?

Final Guidlines for inter-hospital transport of critically ill patients

Diagnosis

- Patient requires emergent surgical intervention that will occur immediately upon arrival to the accepting institution. This applies to diagnoses such as cerebellar hematoma, EDH, or SDH in the setting of a rapidly deteriorating patient. This defines the highest acuity transfer wherein communication between physicians, OR staff, and transport personnel is critical to transporting a patient directly to the holding area and/or operating room in order to save the patient's life. Expending time to "make a bed" at the accepting institution will gravely impact the patient's outcome and thus must be overlooked in order to transport the patient as rapidly as possible to the operating suite.
- **Patient requires emergent surgical intervention within 24-hours of arriving to the accepting institution.** This applies to diagnoses such as SAH where cerebral angiogram with definitive treatment is performed within 24-hours of arrival. The caveat here is that these patients may develop hydrocephalus which sometimes cannot be treated at a referring institution and therefore requires emergent transport. Another diagnosis is cauda equina where a patient will require an MRI followed by surgery within 24-hours.

- **Patient requires urgent imaging studies that may or may not require surgery in the setting of a neurologically and medically stable patient.** This applies to diagnoses such as cervical spine trauma where the patient's neurological status is not changing. For example, a patient with facet dislocation requiring traction and/or reduction.
- **Patient requires observation in a neurosurgical ICU, but remains stable neurologically.** This applies to diagnoses such as a basal ganglia hemorrhage.
- **Patient does not have an established diagnosis due to lack of MRI, angiography, etc. at the referring institution but remains neurologically stable.**

PRE-TRANSPORT COORDINATION

Communication is Key

- Establish continuity of care by physician-to-physician and nurse-to-nurse review of patient condition and current treatment plan. Receiving physician should provide advice to aid in pretransport stabilization and provide advance medical treatment when appropriate (ie, Mannitol, steroids, antiepileptic in setting of aneurysmal SAH with hydrocephalus). The appropriate arrangements should be made at the receiving institution to avoid delays in definitive treatment (ie, operating room standby). The mode of transport must also be established. Communication is especially critical in the setting of the first set of diagnoses described above.

TRANSPORT PERSONNEL

Education = Empowerment to Intervene

- A minimum of two people should accompany a critically ill patient consisting of a critical care nurse plus a technician. In the case of an unstable patient, it is recommended that a physician with training in airway management and ACLS be present. When this is not possible, some hospitals prophylactically intubate those patients who have a high risk for becoming unstable en route. In the setting of elevated ICP, the transport personnel must understand the diagnosis and means of controlling ICP during transport. For example, most patients arrive flat and supine which is not the optimal position for a patient with impending hydrocephalus. Transport personnel cannot serve as technicians, but must be aware of the patients diagnosis in order to best maintain and even optimize the patient prior to arrival.

TRANSPORT EQUIPMENT

Continuity of Care

- The patient must be maintained using the same monitors and drugs (if necessary) as are present in the ICU. A blood pressure monitor, pulse oximeter, cardiac monitor/ defibrillator, basic rescucitation drugs, and a portable mechanical ventilator when indicated.

DOCUMENTATION:

What is the Goal of Transfer?

- The patient's medical record and relevant laboratory and radiographic studies must be copied for the accepting facility. It is also suggested to perform a COBRA/EMTALA checklist to ensure compliance with federal regulations. This includes documentation of initial medical evaluation and stabilization procedures, informed consent disclosing risks and benefits of transfer, and documentation of physician-to-physician communication with the names of each physician involved. Financially motivated transfers are illegal.

The above five points serve as a general set of guidelines that should be evaluated and refined regularly by the hospital using a standard quality improvement process. The hospital transfer center should develop a checklist for each of the five points described above and follow the same steps for each and every transfer. Attention to details by methodically following the same steps in each situation will hopefully minimize "systems" mistakes and thus not only improve efficiency, but also maximize safety.

Spinal Cord Stimulators: an Introduction

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Background

Pain can be divided into two broad categories, nociceptive pain and neuropathic pain. Nociceptive pain is a dull, throbbing pain which results from irritated nerves after physical tissue injury. This is seen commonly in cancer or after a fracture. Nociceptive pain is amenable to treatment with pain medications such as opioids and/or anti-inflammatories. Neuropathic pain is described as burning, shooting, or shocking pain. This type of pain results from nerve damage or abnormal nerve conduction such as pain exhibited with failed back syndrome, post surgical pain, neuromas, shingles, and complex regional pain syndrome (previously called RSD or causalgia). Neuropathic pain tends to be resistant to treatment with pain medications. Neurostimulation has been an effective treatment option for the management of chronic neuropathic pain. It is a reversible therapy which can even be tested before permanent implantation.

Spinal cord stimulation (SCS) is an adjustable, non-destructive, neuromodulatory procedure which delivers therapeutic doses of electrical current to the spinal cord or to a targeted nerve. This low-voltage stimulation can block the transmission of pain. The enthusiasm for SCS began with the introduction of the gate control theory for pain control by Melzack and Wall in 1965[1].They noted that stimulation of large myelinated fibers of peripheral nerves resulted in paresthesias and blocked the activity in small nociceptive projections. In other words, pain receptors compete with each other and with other sensory afferents. Appropriate stimulation of a "rival" afferent can effectively block a pain signal. This is why rubbing your chin after its been hit relieves the pain – the bump is still present, but the rubbing blocks it. The SCS system is implanted in a space surrounding the spinal cord, called the epidural space, where it stimulates the dorsal columns which can mask the sensation of pain by producing a tingling sensation.

Patient Selection

Some of the criteria listed below can be used to determine candidates for neurostimulation:

- Conservative therapies have failed to adequately help the pain.
- Further traditional surgical intervention is not indicated.
- No serious untreated drug habituation for the pain condition exists.
- Psychological evaluation and clearance for implantation have been received, sorting out untreated depression or anxiety, addiction, or other behavioral disorders.
- No medical issues exist that would present problems with doing the surgery.

Indications

The most common indications include post-laminectomy syndrome or failed back surgery syndrome (FBSS), complex regional pain syndrome (CRPS) or reflex sympathetic dystrophy (RSD), ischemic limb pain, and angina. Scattered reports regarding the treatment of intractable pain due to other causes including visceral/abdominal pain, cervical neuritis pain, spinal cord injury pain, post-herpetic neuralgia, and neurogenic thoracic outlet syndrome have also appeared in the literature.

Surgical Technique

The procedures are most commonly performed by neurosurgeons or anesthesiologists specializing in pain management[2-5]. A lead is temporarily utilized to cover a wide area for pain control. The lead, or electrode, is implanted into the epidural space. The lead is attached to a power source called the pulse generator (or commonly known as the battery). This is then connected to an outside or external system which is held in place by sterile dressings. The unit is then tested for efficacy during a screening period, which is usually 3-10 days.

Figure 1

A patient that had persistent neuropathic pain in his back and his legs despite anterior and posterior spinal reconstructive surgery.

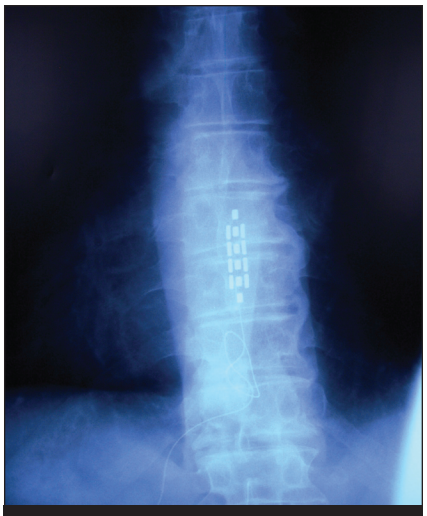

Figure 2

The patient received an implanted transverse tripole electrode in the thoracic spine. The new electrode has the ability of focusing the electrical field into the dorsal column to maximize the paresthesia into the back while avoiding painful stimulation of the nerves into the ribs.

If relief is obtained at this point, the system may be implanted permanently. The stimulator is usually placed in an area where the skin will not be irritated and the unit will not be pressing against bone. The abdomen and buttocks are common locations.

Benefits

Neurostimulation has been shown to have a 50% improvement in pain relief, reduce the use of medications, and allow the person experience an improvement in their quality of life [6-7]. These benefits are expected to increase with improvements in the technology and in understanding of how pain signals are generated and gated in the nervous system. It is important to realize that neurostimulation is a treatment option along the continuum of pain control.

Potential Adverse Effects

Surgical risks associated with an implant include hematoma, infection, and possible leakage of cerebrospinal fluid. Because this is a device, there can be malfunction of the hardware or can have disruption in the settings by other signals; for example, patients with an SCS are advised to turn it off before going through an anti-theft device in a retail store; additionally, patients with an SCS can not undergo a magnetic resonance imaging (MRI) scan.

Summary

Spinal cord stimulation therapy has been available and approved for over two decades. In the last five years, there have been significant advances in the technology: smaller, rechargeable batteries; better electrodes; and improved implantation techniques. More physicians are adapting this therapy in their practice for pain management and we are effectively improving patients' lives.

The treatment of chronic pain remains challenging. Experience in the technique and the equipment has made SCS a much more reliable and safe modality. Like all the modalities performed for chronic pain management, its results are favorable. It is important to remember that the goal of neurostimulation is to reduce pain, rather than to eliminate pain. Very few other invasive modalities can claim this success rate with a few years of follow-up. Spinal cord stimulation has earned a well established and firm role in contemporary chronic pain management.

Further information is available at the Neural Interfaces Program Website of the NIH, *http://www.ninds.nih.gov/funding/research/npp/*

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Surgeon Opinions on Use of Epidural Steroids in Treatment of Lumbar Disk Disease: Results of an Online Survey

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Introduction

"Standard of care" can vary along regional and specialty lines; it is common to discover that a local "standard" can be different somewhere else. Opinions may differ between pain management specialists, primary care physicians, and spine surgeons with regard to use of conservative treatment modalities.

Opinion within a given group of practitioners, however, should converge. Local differences between hospitals may exist, but conferences, professional journals, and national boards for certification are mechanisms that should act to maintain homogeneity within a professional group. It could be expected that commonly utilized treatment approaches within a well defined group of sub-specialists should converge.

One of the more common non-surgical options for herniated lumbar spinal discs is epidural steroid injections (ESIs). Patients may be referred to pain management centers for lumbar ESIs by their primary care physician or perhaps after consultation with a surgical specialist. We sought to assess the opinion of practicing spine surgeons with regard to timing and use of lumbar ESIs as a part of a conservative treatment approach to both lumbar disk herniations (HNP) and lumbar degenerative disk disease (DDD).

Table 1.

1. When in the treatment plan do you consider ESIs for patients with a lumbar herniated disc?	a. after 6 weeks of conservative care b. after 6-12 weeks of conservative care c. after 3-6 months of conservative care d. after greater than 6 months of conservative care
2. What is the average number of ESI treatment cycles that patients with lumbar herniated discs normally receive before considering alternate treatments?	a. 1 treatment regimen b. 2 treatment regimens c. 3 treatment regimens d. 4 treatment regimens or more
3. How long are patients with lumbar herniated discs treated with conservative care and ESIs before more invasive treatment options are considered?	a. up to 6 weeks b. 6-12 weeks c. 3-6 months d. Patient directed
4. How frequently do you consider using epidural steroid injections (ESIs) in your treatment of patients with lumbar herniated disc?	a. Frequently (>75% of the time) b. Often (50-75% of the time) c. Sometimes (25-50% of the time) d. Rarely (<25% of the time)
5. How many ESIs are normally considered in a treatment regimen for patients with lumbar herniated discs?	a. 1 injection b. 2 injections c. 3 injections

The definition of "conservative therapy" is unclear, although use of same as a control arm in prospective studies of surgical efficacy is commonplace. We sought to assess whether practicing spine surgeons would agree upon treatment protocols in their approach to treatment of lumbar degenerative disease and lumbar disc herniations. Lack of agreement on use of this tool in conservative management of lumbar disease could imply greater divergence in broader use of conservative treatment modalities.

Methods

An online survey was announced and made available on www.spineuniverse.com, a portal site for information on therapies for spinal conditions. Sixty-one surgeons responded to the survey. Because participants were self-selected, we must be cautious about whether the results can be generalized. The survey questions are presented in Table I. Sixty-one surgeons responded.

There was broad divergence in all responses. The only clear majority was the decision to consider ESIs after 6 weeks of conservative treatment in lumbar HNP (Figure 1). In both HNP and DDD, the number of ESI regimens administered before considering surgical alternatives followed the same pattern: surgeons were likelier to favor smaller numbers of cycles, and going as far as 3 or 4 regimens was unlikely (Figure 2). Otherwise, there was no clear pattern in how ESIs were used (Figures 3, 4 and 5).

Bowker's test of symmetry was used to assess whether surgeons were likely to use ESIs the same way in HNP and DDD. Statistically significant differences were found in the length of time before ESIs were considered (Figure 1; $p < 0.01$) and the length of time before invasive surgical options were considered (Figure 3; $p < 0.05$). These results suggest that surgeons tend to change treatment options earlier in HNP than in DDD.

Discussion

If the self-selected response sample is biased, we might assume that the bias would tend to homogenize the reported opinions. Conversely, a small population sample may

Survey Responses

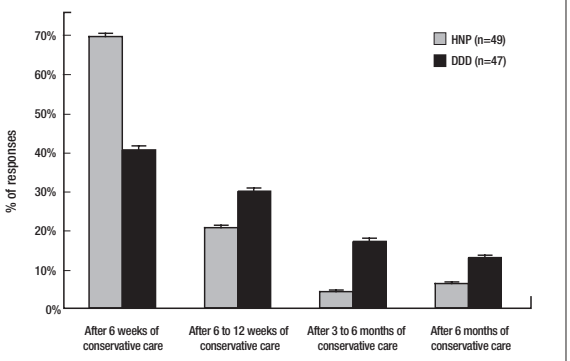

Figure 1
When in the treatment plan do you consider ESIs for patients with a lumbar herniated disc?

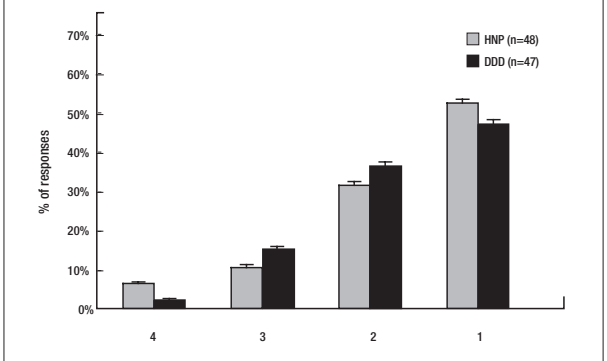

Figure 2
What is the average number of ESI treatment cycles that patients with lumbar herniated discs normally receive before considering alternate treatments?

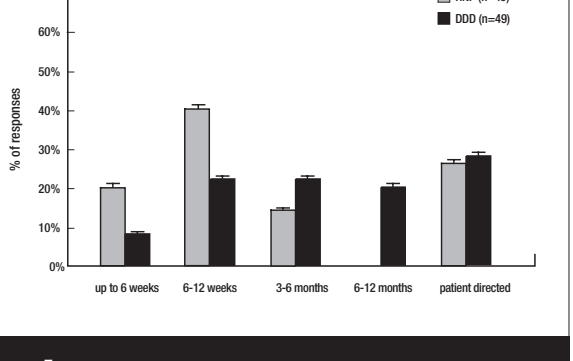

Figure 3
How long are patients with lumbar herniated discs treated with conservative care and ESIs before more invasive treatment options are considered?

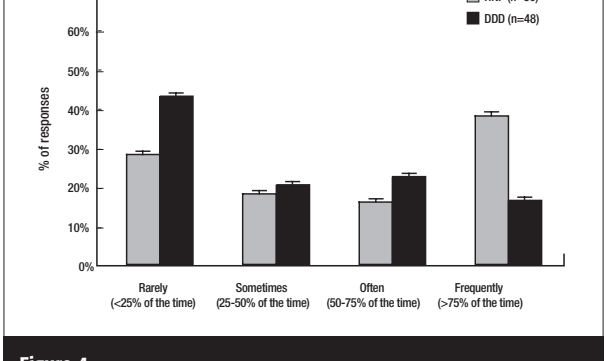

Figure 4
How frequently do you consider using epidural steroid injections (ESIs) in your treatment of patients with lumbar herniated disc?

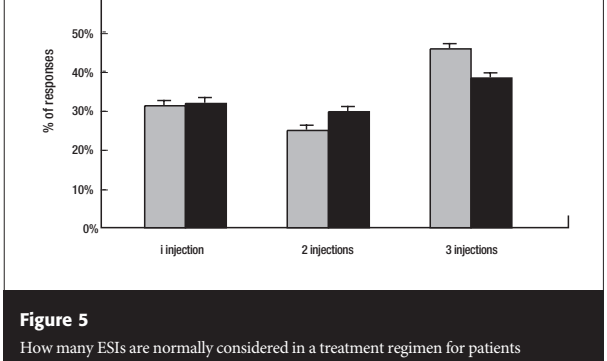

Figure 5
How many ESIs are normally considered in a treatment regimen for patients with lumbar herniated discs?

show more difference from the "true" mean than would a larger sample. With these caveats, we may state based upon our survey that there is no clear consensus on how the role of epidural steroids in treatment of patients with HNP or DDD.

The potential consequence on patient care is obvious. Treatment regimens given to individual patients are effectively randomized according to which surgeon assesses their case. While there may be no significant difference on outcome between three weeks or six months of ESI treatment, there remains a potential real impact on patient well-being and on medical expenses incurred.

More importantly, this variance may also complicate attempts to study the effectiveness of new interventions. If "conservative" control treatments vary from surgeon to surgeon, and presumably from location to location, studies using "standard conservative treatment" as their control arm are suspect. It is possible, though untestable, that these variations self-propagate as surgeons base their choices on both published articles and on communication between colleagues.

Case Report: Intramedullary Cervical Spinal Cord Hemangioblastoma with an Evaluation of von Hippel-Lindau Disease

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History of Present Illness

MO is a 49 year old male with a history of multiple sclerosis who presents with a one year history of progressive numbness in his shoulders bilateral and upper back. The patient describes occasional sharp pains that radiate to his first three fingers on his right hand. He denies weakness, clumsiness, difficulty walking, or bladder/bowel dysfunction. He describes no problems with handwriting, or fine motor skills.

Past Medical History	Hypertension, Diverticulits, Multiple Sclerosis, optic neuritis
Past Surgical History	Inguinal hernia repair, Colectomy, Re-anastomosis of bowel
Social History	Inguinal hernia repair, Colectomy, Re-anastomosis of bowel
Allergies	NKDA
Medications	Toprol, Beta Interferon
Initial Neurological Exam	The patient's exam was consistent with a decrease to light touch in his bilateral lower extremities and right upper extremity. Pinprick and proprioception were intact. Motor strength was 5/5 in all muscle groups. There were no difficulties with tandem gait. Reflexes were 2+ throughout and symmetric. The patient had showed neither Babinski's nor Hoffman's sign.

Hospital Course

A MRI of the brain was performed early in the course of symptoms demonstrating his known multiple sclerosis lesions without any changes. With continued worsening symptoms, further workup included an MRI of the cervical spine which demonstrated a 1 cm homogenously enhancing intramedullary lesion at C2 with an associated cyst and syrinx.

Prior to surgical intervention, the patient was counseled on the risks entailed in the removal of a spinal cord tumor. Based on the characteristics on the MRI, a diagnosis of hemangioblastoma was favored and therefore the possibility of having von Hippel-Lindau (VHL) disease was discussed with the patient. The patient underwent an MRI of the complete neuro-axis including brain and spinal cord screening for any other lesions. An ultrasound of the abdomen was performed which demonstrated no renal lesions, liver lesions, or adrenal masses. An ophthalmologic examination demonstrated none of the stigmatisms associated with VHL.

Prior to surgical excision, an angiogram was performed for the potential to embolize any large vessels. However, no tumor blush was appreciated during the angiogram.

The patient underwent a laminectomy from C1-3 to expose the spinal cord. Intra-operative ultrasound was used to isolate the borders of the nodule and define the cystic component. A dural incision and midline myelotomy was performed. The tumor was removed en bloc and the cyst decompressed. The tumor was noted to be significantly vascular. Intra- operative ultrasound was utilized to confirm full resection. During removal of the tumor somatosensory evoked potentials were noted to decrease by approximately 50% in all four extremities.

The patient worked with physical therapy and was discharged home on post operative day number four. His post operative incisional pain was controlled with Percocet. The patient was also administered Valium and Neurontin to help with the neck pain and parasthesias following the surgery. The patient's neurological exam on discharge demonstrated mild numbness in the right upper extremity and slight weakness in right hand grip, with resolution of preoperative sensory deficits in the lower extremities. Motor exam was 5/5 in all other muscle groups. Postoperative MRI demonstrated no residual tumor with expected post operative changes. Pathology was consistent with hemangioblastoma. In follow up the patients remaining right upper extremity numbness had resolved and the patient was able to return to work 8 weeks postoperatively.

Discussion

The decision to proceed with surgery was based on the natural history of this tumor. It may continue to grow and with further expanding of the syrinx or cystic cavity may produce further neurological deterioration. It can be anticipated that the best time for surgery would be while the patient has a strong neurological status and best chance for recovery.

Hemangioblastoma is a vascular neoplasm of the central nervous system. It is the most common primary neoplasm in the adult cerebellum, but is rarely a tumor of the spine[1]. Hemangioblastoma may occur sporadically or as a component of von Hippel-Lindau syndrome. Radiological findings such as associated cyst, an associated syrinx, and/or surrounding spinal cord reaction or edema[2, 3] are helpful in diagnosing hemangioblastoma. Thirty-two percent of patients with spinal hemangioblastoma have von Hippel-Lindau syndrome. Symptoms of spinal hemangioblastoma are similar to those of other spinal canal tumors and include sensory change, motor disturbance, and pain. Because the presentation of spinal hemangioblastoma is similar to that of other spinal canal masses, MRI studies can be useful in suggesting a diagnosis[2].

VHL is a rare genetic condition. The disease is an autosomal dominant disorder carried on the third chromosome with near complete penetration and variable expression[4]. The gene behaves

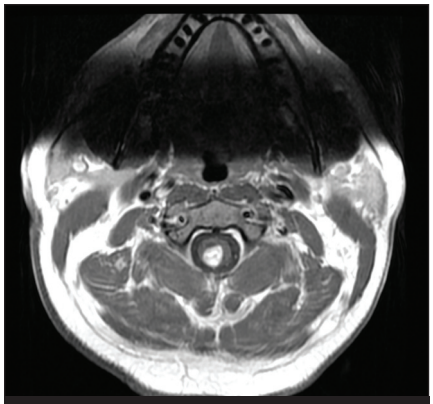

Figure 1
T1 axial MRI post contrast

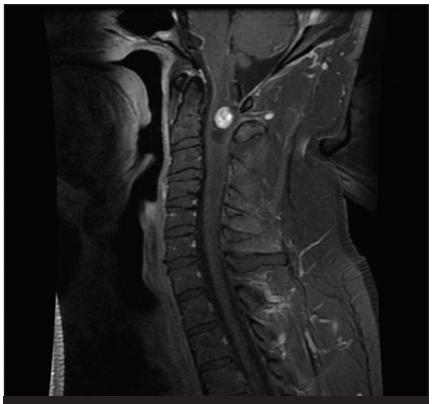

Figure 2
T1 sagittal MRI post contrast

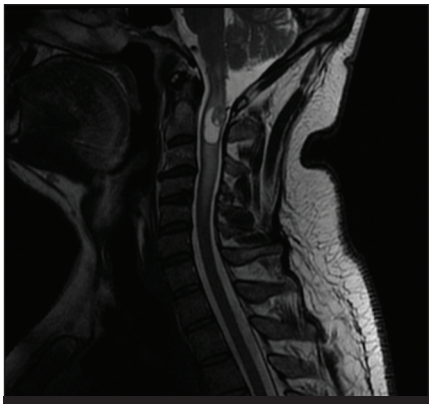

Figure 3
T2 sagittal MRI

as a typical tumor suppressor gene. The age of onset, as well as severity is variable. There are various subtypes which characterize the risks of developing renal call carcinoma, pheochromocytoma, and angiomatosis. Angiomas are the most common presenting signs and symptoms of this disease syndrome. Of note, only 20% of patients with VHL develop pheochromocytomas.

Features of VHL can include hemangioblastoma's in the cerebellum and spinal cord, as well as angiomatosis in the retina and other organs.[1] Other features include pheochromocytoma, renal cell and pancreatic cysts, renal cell carcinoma, as well as café au lait spots. Neurological sequeala and death arise from complications associated with central nervous system tumors, as well as cardiovascular instability secondary to a pheochromocytoma. Decrease in visual acuity and blindness may arise as well[5].

Work up for suspected VHL can include genetic studies, as well as Vanillylmandelic acid levels in urine. Imaging studies can include an MRI of the brain and spinal cord, ultrasound and/or CT of the abdomen, and ophthalmic exam with ocular dopplers[5]. VHL is usually a progressive disease and therapy should begin as soon as the diagnosis is made. Surgical treatment is usually indicated.

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Cervical Intramedullary Ganglioma

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A 48 year male presented to the ER with severe headaches which were episodic in nature and which had been present for several weeks. Patient had a history of traumatic head injury (TBI) several years prior. Otherwise, he was in good health with no significant past medical or surgical history.

On physical exam, patient was oriented x 3 with an intact cranial nerve exam. He had significant upper and lower extremity spasticity with mild hand intrinsic weakness. His motor exam was otherwise unremarkable. His gait was very spastic. He had sustained lower extremity clonus, upgoing toes, and increased tone in the upper and lower extremities. His sensation was intact to light touch, pinprick, proprioception and temperature.

What imaging studies would you order?

A CT scan of the brain was initially obtained secondary to the severity of the patients' headaches and past history of TBI.

The head CT revealed an old lacunar infarct in the left basal ganglia. There was no intracerebral hemorrhage or extra axial collection. However, in the spinal cord at the level of C1, an 8 mm hypodensity was noted.

This prompted an MRI of the cervicomedullary junction. An ovoid, 1.1 x 1.0 x 1.7 cm intramedullary lesion was seen expanding the cervical cord at the C1-2 level. It was hyperintense on T2-weighted images and hypointense on T1 weighted images, and demonstrated some mild curvilinear enhancement within the lesion.

What is your differential diagnosis? Would you perform any further imaging?

These radiographic findings were consistent with a low grade tumor such as an astrocytoma. An ependymoma or cord metastasis were considered less likely.

No evidence for primary malignancy was found on imaging of the chest, abdomen and pelvis.

Imaging of the entire neural-axis did not reveal any other lesions.

A CSF flow study on MRI was also performed to look at CSF flow dynamics, and evaluate for possible CSF outflow obstruction secondary to this lesion, which might have explained the patient's headaches. Unobstructed CSF flow was identified anterior and posterior to the cervicomedullary junction at the foramen magnum and at the level of the intramedullary lesion at C1-C2.

How would you treat this lesion?

A. Observation and close neurological follow up

B. Perform a lumbar puncture for CSF cytology

C. Radiation and/or chemotherapy

D. Excisional biopsy

E. Excisional biopsy followed by radiation and/or chemotherapy

This is a young, otherwise healthy male who is starting to develop neurological symptoms from a lesion with unknown pathology. Considering the rapid progression of his symptoms over a few weeks, we felt that it was necessary to expediently secure him a diagnosis via direct exploration of the lesion. Once the diagnosis was established, treatment tailored to his particular ailment could begin. While operative exploration of this lesion entails risk of significant morbidity, we felt the severity of his clinical findings and the progression of his deficit merited operative exploration.

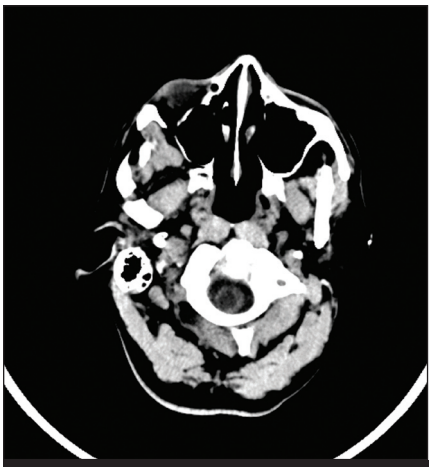

Figure 1

Pre-op axial non-contrast CT head showing hypodensity within the spinal cord

Figure 2

Pre-op sagittal T1-weighted MRI

The diagnostic yield from CSF cytology is low (~26%)(3), although repeated sampling does increase the yield significantly. CSF flow cytometry also increases the diagnostic yield over CSF cytomorphology alone. (1) Newer techniques like fluorescence in situ hybridization (FISH) enhance the diagnostic yield for

Figure 3
Pre-op sagittal T2-weighted MRI

Figure 4
Pre-op sagittal post-gadolinium MRI

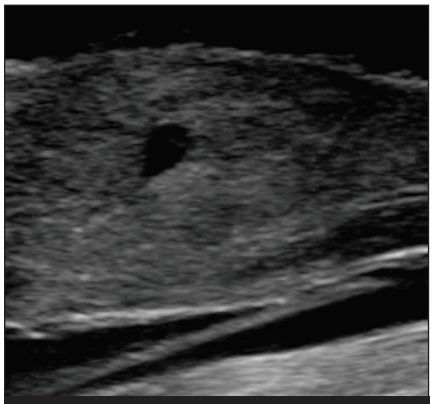

Figure 5
Intraoperative ultrasound image, pre resection

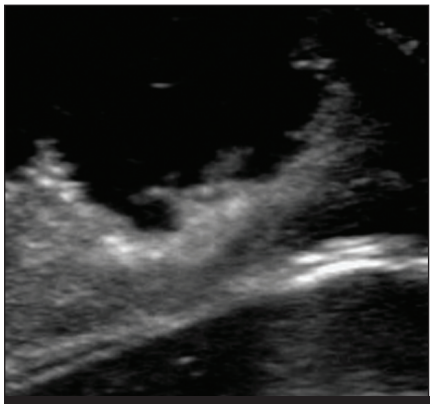

Figure 6
Intraoperative ultrasound image, post resection

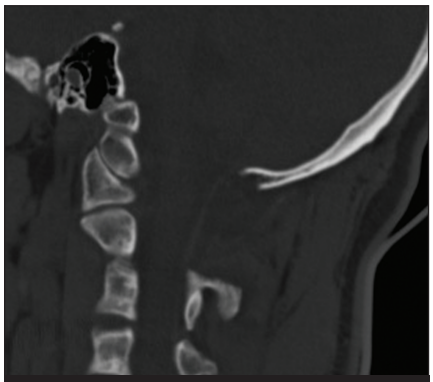

Figure 7
Post-operative sagittal CT

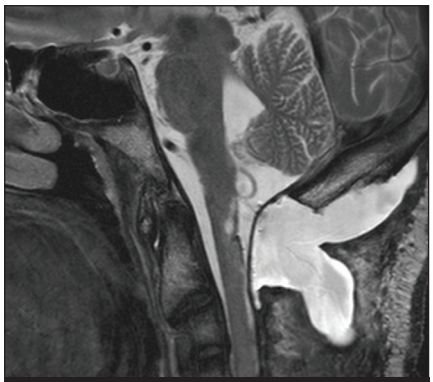

Figure 8
Post-operative sagittal MRI

the detection of malignancy on the first lumbar puncture in patients clinically suspected of having leptomeningeal metastases (LMM). (10) Radiographic imaging did not demonstrate any leptomeningeal spread in our patient. In the absence of leptomeningeal spread, the chances of obtaining a diagnosis from CSF cytology are further diminished.

We decided to proceed with exploration and biopsy of the tumor. We planned to debulk the tumor and to possibly resect the entirety of the tumor predicated upon the lesion's histopathological findings at the time of frozen section and intraoperative behavior. Debulking the tumor would also alleviate the mass effect on the spinal cord, decrease tumor burden, and afford the patient a better chance at tumor control through adjuvant chemotherapy and radiation.

How would you counsel the patient regarding the surgical risks?

The risks and benefits of surgery were explained at length. It was emphasized to the patient that there was a substantial risk of neurologic worsening and a significant risk of paralysis secondary to the size of the tumor and its extremely sensitive location. The patient had previously suffered a significant head injury; hence, these discussions were also carried out with patient's mother who acted as his guardian. While the patient was felt to be thoroughly consentable, the patient's family was again counseled as to the severity of his diagnosis and the significant dangers of surgery.

What surgical approach would you use? What intra-operative tools would help you in localization of this lesion?

We decided to approach this lesion through a posterior cervical approach. The patient was positioned prone using the Mayfield three-pin headholder. The patient's neck was placed in gentle flexion to facilitate exposure of the occipitocervical junction. The posterior arch of C1, along with 50% of the top of the lamina of C2 was removed. At the base of the occiput, we performed a limited suboccipital craniectomy using the Midas-Rex drill.

The dura was opened next. Using intraoperative ultrasound localization, we identified the patient's large intramedullary tumor. We attempted to map the spinal cords' midline using SSEPs; however, due to poor recordings and extensive lateral spread, we were unable to adequately map the posterior elements.

We chose an entry point along the posterior elements. The spinal cord was entered via bipolar and creating a sharp incision in the

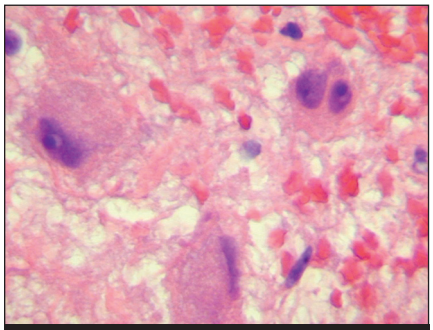

Figure 9
Atypical ganglion cells including binucleated ganglion cell

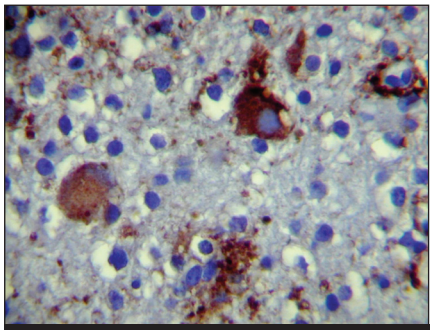

Figure 10
Chromogranin staining of atypical ganglion cells in background of neoplastic glial cells

pia mater. The cord was opened further using microscissors. Using blunt dissection, we gently reflected the pia-arachnoid. Using pial sutures, we gently retracted the spinal cord bilaterally. The patient's intramedullary tumor was evident. Using blunt dissection, we gently developed a plane around the patient's intramedullary tumor. A biopsy was obtained and sent for frozen section. Frozen section returned a low-grade astrocytoma.

We continued our resection, removing as much of the tumor as we felt safe. We periodically checked motor evoked potentials during our decompression. After we had completed our decompression, we used the ultrasound to confirm that no further gross elements of tumor were evident. There was a gliotic shell, which was evident around the spinal cord; however, based on ultrasound localization, the main portion of the patient's tumor had been successfully extirpated.

There was some mild loss of hand intrinsic motor evoked potentials during the procedure; however, this was less than 50% loss and was not felt to be severe. After hemostasis had been achieved, the wound was thoroughly irrigated. The dura was closed using #6-0 Gore-Tex sutures.

Postoperatively, the patient recovered well. His motor and sensory exam remained stable.

Permanent section was consistent with that of ganglioglioma.

Ganglioglioma was first described by Courville in 1930 as a central nervous system neoplasm containing both astrocytic and neuronal components.

Gangliogliomas are firm grayish tumors that may have cystic components. On light microscopy, atypical dysplastic neurons, astrocytes, and fibrovascular stroma are observed. Infrequently, anaplastic degeneration occurs and involves the astrocytic component. Neoplastic ganglion cells are large and mature and reveal intense immunoreactivity with synaptophysin, show expression of the stem cell epitope CD34, and are often binucleated.

Most gangliogliomas are observed in the brain. Temporal lobes and cerebellar hemispheres are the most common locations. Rarely, they may also develop in the spinal cord. Only 1% of intramedullary spinal neoplasms are histologically gangliogliomas.

Now that diagnosis has been established, would you recommend any further treatment for this patient?

Radical surgery for intraspinal tumors can achieve long tumor-free survival without requiring adjuvant therapy.(4) In cases of low-grade tumor, radical excision is associated with minimal morbidity and an excellent long-term prognosis when carried out before significant disability occurs.(2) Gangliogliomas are generally classified as WHO grade 1 lesions; however, malignant transformation can occur.

In one study, the 5-year actuarial survival rates for cerebral hemisphere, spinal cord, and brain-stem gangliogliomas were 93%, 84%, and 73%, respectively ($p = 0.7$). The event-free survival rate at 5 years was 95% for cerebral hemisphere gangliogliomas and 36% for spinal cord gangliogliomas ($p < 0.05$); for brain-stem gangliogliomas the event-free survival rate at 3 years was 53% ($p < 0.05$). Multivariate analysis (Cox linear regression) revealed tumor location to be the only variable predictive of outcome, with spinal cord and brain-stem gangliogliomas having a 3.5- and 5-fold increased relative risk of recurrence, respectively, compared to cerebral hemisphere gangliogliomas. Histological grade was not predictive of outcome, although in each location there was a trend for higher-grade tumors to have a shorter time to recurrence.(6)

It is concluded that radical surgery leads to long-term survival of patients with gangliogliomas, regardless of location, and adjuvant therapy can probably be reserved for special cases (e.g. tumors with a high KI 67 index). (4, 5, 7-9)

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Intramedullary Spinal Cord Metastases and Radiation Therapy: A Case Report

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Introduction

Intramedullary spinal cord metastases (ISCM) are a clinically rare, although devastating, complication of disseminated cancer. These lesions have been reported to originate from many types of solid tumors, although primary lung carcinoma, particularly small cell, is the most common etiology. These metastases, which can occur anywhere along the spinal cord, often represent the end-stage of the disease process with limited survival outcomes.

Patients with ISCM may develop a variety of neurological deficits with treatment goals aimed at palliation. Different modalities of treatment have been found to preserve or restore ambulation and neurological function. The options for therapeutic intervention include surgical, chemotherapeutic, and radiation therapy. We describe a case of ISCM in a patient with disseminated small cell lung cancer with magnetic resonance imaging that illustrates a complete tumor response to radiation therapy

Case Report

In December of 2006, a 63 year-old male presented with extensive stage small cell lung carcinoma. Upon initial presentation, this patient had a very large, left pulmonary mass, mediastinal lymphadenopathy, multiple hepatic lesions, and several osseous lesions. Due to the extent of his disease, the patient was referred to medical oncology and treated with chemotherapy. A PET scan on July of 2007 revealed radiographic resolution of his malignancy following his chemotherapy. On October 3rd of 2007, after presenting with dizziness, it was found he had metastasis to his cerebellar vermis. These metastases were treated with 30 Gy of whole brain radiation therapy and his vertigo significantly improved. A follow-up brain MRI showed a complete response to treatment with no residual metastases found.

In December of 2007, the patient presented with bilateral shoulder and neck pain. The pain started in his right shoulder radiating to his mid neck and then to his left shoulder. The pain had been intermittent, but intense, over the previous few weeks and progressed to right arm numbness. There was no associated muscle weakness in his upper or lower extremities bilaterally. The patient displayed some mild right hand intrinsic weakness; deep tendon reflexes were found to be slightly increased in his lower extremities versus his upper extremities. An open MRI with and without IV contrast of the cervical spine was performed. A contrast enhancing mass within the cervical spinal cord was noted and reported as an ependymoma, due to the infrequency of intraparenchymal metastasis. To get better definition of the lesion, a repeat closed high-resolution MRI of the cervical, thoracic, and lumbar spine was performed. The cervical contrast enhancing lesion was found to be three separate intramedullary multifocal with measured total length of 4.5 cm (Figure 1A). There were no other lesions in the spinal cord. The presence of three distinct lesions in the setting of metastatic disease was consistent with multiple intraparenchymal metastatic disease rather than a primary intramedullary neoplasm (ependymoma).

The patient's disease was in remission and 30 Gy of cervical spine radiation was administered. The patient reported that his symptoms of pain and numbness resolved completely with the radiation therapy.

A follow-up MRI demonstrated a complete response and no evidence of the ISCM lesion (Figure 1B).

Discussion

Although rarely reported, ISCM have been found in 0.9% to 2.1% of cancer patients by autopsy, suggesting that thousands of patients are afflicted yearly[1]. Primary lung carcinoma is the most common etiology and reported to occur in up to fifty percent of cases of ISCM[2]. In one retrospective study, small cell lung cancer was the most common primary tumor in patients with ISCM, accounting for 63% of patients with primary lung cancer and 30% of all patients studied[2]. However, other solid tumors, including breast, kidney, colorectal, cervical, and ovarian cancers have also been reported[3,4,5].

A variety of neurological disorders characterizes the presenting symptoms of ISCM clinically. Schiff and O'Neill reported sensory alteration (42.5%), pain (30%), and weakness (30%) as the most common initial symptoms[2].

Figure 1A
Cervical MRI, prior to 30 Gy radiotherapy

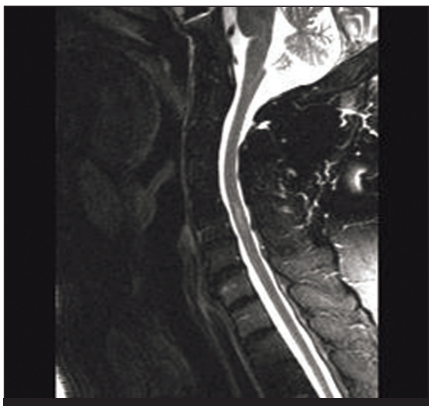

Figure 1B
Cervical MRI, post radiotherap

The symptoms of gait unsteadiness, urinary incontinence, and Brown-Sequard syndrome have also been described[2,6].

Diagnosis of ISCM usually requires magnetic resonance imaging (MRI) or computerized tomography (CT), because plane films and myelography are normal in 75% and 58% of cases[7]. In addition, cerebral spinal fluid (CSF) analysis is often non-diagnostic with the most common abnormal finding of elevated protein in only 20% of cases[8]. Computerize tomography scanning has been found to be diagnostic in few patients, which is demonstrated by increased density in the area of the ISCM lesions. However, magnetic resonance imaging (MRI) has greater sensitivity and specificity[7]. MRI T2-weighted images show alterations in signal intensity based on tissue type and demonstrate accurate differentiation between normal tissue of the spinal cord and tumor[7]. Gadolinium is a helpful adjunct to the T2-weighted imaging as it reveals the enhancing central lesion in typical ISCM[7]. In a study of 30 patients, there was only one false-negative MRI study, and 23 of 25 patients receiving gadolinium demonstrated contrast enhancement of ISCM lesions on MRI[2].

Treatment is generally palliative, as the median survival ranges from 3.9 to 5 months[2,9,10]. Schiff and O'Neill found that patients with breast cancer as their primary tumor had the greatest median survival, which still was only 13 months. Patients with other primary cancers including lung carcinoma had a median survival of 3 months. As with our patient, surgery is often contraindicated at the time of presentation, making the less invasive modalities of chemotherapy and radiation therapy the mainstays of treatment[2,5,11]. Radiation therapy has been shown to improve or prevent deterioration of neurological status. In Conill et al., they reported improvement of neurological symptoms in 83% of patients with a mean duration of 17.2 days. And Schiff and O'Neill reported preservation of ambulation in 91% of patients at their latest follow-up.

Conclusions

This case illustrates a rare complication of systemic small cell lung cancer. The ISCM lesion showed a rare complete response to radiation therapy, which was evidenced by magnetic resonance imaging and outlines the importance of radiation therapy in the treatment algorithm of these patients.

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Research
Department of Neurosurgery

Clinical Research Ongoing Studies

- Survey on Whether Patients Will Participate in a Blood Transfusion Trial after Subarachnoid Hemorrhage
- Retrospective, Multi-Center Chart Review of the Wingspan™ Stent System with Gateway™ PTA Balloon System.
- Outcome of Patients with Ruptured Aneurysm Who Underwent Endovascular Coiling Procedure and Evacuation of Intracerebral Clot
- Micrus® Cerecyte™ Registry: a Registry to Evaluate Outcomes at One Year Post-Embolization with the Micrus® Cerecyte™
- A Randomized, Clinical Trial of Unruptured Brain Arteriovenous Malformations (ARUBA)
- Carotid Revascularization Endarterectomy vs. Stent Trial (CREST)
- Surgical Anastomosis of the Superficial Temporal Artery to the Middle Cerebral Artery (STA-MCA) when Added to the Best Medical Therapy for Carotid Occlusion (COSS)
- Assessment of Intracranial Aneurysm Shape as an Indicator of Rupture Risk
- Retrospective 5 Year Review of Follow-Up Angiogram on Patients with Cerebral Aneurysms Post Clipping
- Retrospective Review of Post Treatment Hemorrhage Rates of Patients with Arteriovenous Malformations (AVM's)
- Role of Increased CSF Production in Clinical and Radiologic Vasospasm
- Retrospective and Prospective Review of Peri-Operative Complications in Patients Undergoing Spine Surgery
- Assessing Patient Perspectives of Perioperative Complications in Spinal Surgery
- Retrospective Chart Review: the Use of Pre-Operative and Intra-Operative Ultrasound in the Evaluation of Peripheral Nerve Disorders
- Multicenter Retrospective Study of the Effectiveness, Safety and Treatment Characteristics of Subcutaneous Neurostimulation (SQS) in Patients with Chronic Pain
- Retrospective Review of Surgical Complications of Occipital Nerve Stimulator Systems.
- Comprehensive Retrospective Review of Anterior Corpus Callosotomy Outcomes at TJUH
- Incidence and Outcome of Patients with Spinal Fracture and Ankylosing Spondylitis
- Magnetic Resonance Analysis of Postsurgical Temporal Lobectomy and Correlation to Seizure Outcome
- Study of Seizure Outcomes in Patients Status Post Temporal Lobectomy with Secondarily Generalized Seizures Comparing Patients with and without Intracranial Implants
- Use of Discarded Tissue from Frontal and Temporal Lobectomy
- Pilot Study Comparing Various Electrode and Program Settings Using the EON IPG
- Prospective, Multi-Center, Randomized Controlled Study to Compare the Spinal Sealant System As An Adjunct to Sutured Dural Repair with Standard of Care Methods During Spinal Surgery
- Changing Impedances over Time with Spinal Cord Stimulation Systems
- Comparison of Preoperative ISAP (Intracarotid Sodium Amobarbital Procedure-WADA Test) Results and Post-Operative Outcomes in Patients Undergoing Temporal Lobe Resection for Epilepsy.
- Comparison of Placement of Spinal Cord Stimulators in the Awake Vs. Non Awake Patient
- Pilot Study for the Evaluation of Tripole Electrodes in Spinal Cord Stimulation
- Epilepsy Surgery Complications Relative to Age
- Occipital Cervical Injury Outcomes

- Magnetic Resonance Imaging of Spinal Cord Injury: Correlation with Clinical Findings and Outcome
- Use of Expandable Vs. Non-Expandable Cages in Vertebral Body Replacement Surgery: a Retrospective Review of Outcomes at Jefferson
- Neurological Deterioration After Spinal Cord Injury.
- Radiographic Evaluation of Intersegmental Correction of Cervical Kyphosis with Combined Anterior Interbody Grafting and Posterior Instrumented Arthrodesis
- Magnetic Resonance Imaging of Cervical Spondolytic Myelopathy: Correlation between Findings and Outcome.
- Retrospective Review of the Correlation between Increased T2 Weighted Signal on Cervical Spine MRI to Myelopathic Clinical Signs
- Retrospective Chart Review of Cases Done at Thomas Jefferson University: the Failure Rate in Surgical Treatment with Fixation of C1-C2 Fracture
- Retrospective Chart Review of TJUH Patients Who Obtained Epidural Injections; Looking at the Post Procedural Complications
- Atlanto-Occipital Dislocation (AOD) Injuries: A Retrospective Review of Management and Outcome
- Clinical Outcome in Closed Head Injury: a Retrospective Review of Patients That Received Transfusion (FFP) versus Patients Who Were Not Transfused
- A Research Study of the Surgical Techniques and Clinical Outcome for Interarticularis Pars Repair of the Lumbar Spine
- Clinical Evaluation and Incidence of C 5 Palsy after Cervical Spine Surgery
- Retrospective Review of the Multicenter Spinal Cord Injury Database Utilized for the Pharmacologic Trial Sygen: Defining the Incidence of Neurological Variability after Complete Thoracic ASIA A Spinal Cord Injuries
- Prospective Database of Primary Spinal Neoplasms
- Phase I/II Dose-Ranging Study to Evaluate the Safety, Tolerability, and Pharmacokinetics of BA-210 and the Neurological Status of Patients after a Single, Extradural Application of Cethrin (BA-210) during Surgery for Acute Thoracic and Cervical Spinal Cord Injury (SCI)
- Prospective, Multicenter, Randomize, Study Comparing the Use of HEALOS to Autograft in a Transforminal Lumbar Interbody Fusion (TLIF) Approach
- Single Blind, Randomized Evaluation of Dilute Betadine Solution Irrigation versus Normal Saline Irrigation in the Prevention of Postoperative Posterior Wound Infection in Posterior Instrumented Spinal Surgeries
- North American Clinical Trials Network (NACTN) for Treatment of Spinal Cord Injury
- Flexicore Intervertebral Disc versus Circumferntial Lumbar Spinal Fusion for the Treatment of Discogenic Pain Unresponsive to Conservative Treatment Associated with Degenerative Disc Disease
- Assessment of P-15 Bone Putty in Anterior Cervical Fusion with Instrumentation Investigational Plan
- Multicenter Retrospective Chart Review Reviewing the Safety and Efficacy of the Enterprise Stent in Patients with Intracranial Aneurysm
- Cordis Enterprise™ Vascular Reconstruction Device (VRD) and Delivery System (DS): a Humanitarian Use Device (HUD)
- Humanitarian Use Device: Neuroform Microdelivery Stent
- The Wingspan™ Stent System and Gateway™ PTA Balloon Catheter, a Humanitarian Use Device
- Stenting and Aggressive Medical Management for Preventing Recurrent Stroke in Intracranial Stenosis (SAMMPRIS)
- Pilot Research Study of Coumadin versus Plavix for the Treatment of Phase III, Randomized, Parallel Group, Multi-Centre Study in Recurrent Glioblastoma Patients to Compare the Efficacy of Cediranib [RECENTIN™, AZD2171] Monotherapy and the Combination of Cediranib with Lomustine to the Efficacy of Lomustine Alone
- Use of Fractionated Stereotactic Radiotherapy for the Treatment of Optic Nerve Sheath Meningiomas: a Retrospective Medical Record Review
- Retrospective Chart Review of Fractionated Stereotactic Radiotherapy for the Treatment of Residual Craniopharyngioma
- In Vitro Pilot Study to Determine the Capacity of Glioma Cells and Metastatic Intracranial Tumor Cells to Cross-Prime Dendritic Cells and Induce a Measurable Anti-Tumor Cytotoxic T Cell Response

- Prospective National Study to Molecularly and Genetically Characterize Human Gliomas: the Glioma Molecular Diagnostic Initiative (GMDI)
- Prospective Glioma Patients Archival Database Including Parallel Clinical, Radiographic, Histopathological, and Unsupervised Microarray Analyses
- Exploratory Phase 2 Study Evaluating the Efficacy and Safety of Fibrin Sealant, Vapor Heated, Solvent/Detergent Treated (FS VH S/D) 500 S-APR for the Sealing of Dura Defect Sutures in Posterior Fossa Surgery
- Surgical Treatment of Pituitary Tumor-Transition From Open to Endoscopic Approach
- Overall Experience of Pituitary Tumor Patients at Thomas Jefferson University
- Growth of the Cranial Base Program-Otolaryngology/ Neurosurgery Experience

Neurosurgery Grand Rounds

Overall Goals & Objectives

- Evaluate current controversies in neurosurgery
- Discuss routine occurrences in neurosurgical practice and evaluate them in terms of outcome and alternative methods of management
- Review recent advances and current therapeutic options in the treatment of various neurosurgical disorders.

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For additional information and a schedule of speakers, please contact:
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Fridays, 7:00 am
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